Predictors of quality of life in young adults with congenital heart disease

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Abstract: Aims: The aim of this study was to identify medical and psychosocial risk factors for impaired health-related quality of life (HRQoL) and poor psychological adjustment (PA) in young adults with congenital heart disease (CHD). Methods and Results: A group of 188 patients (43% females, ages 18-30 years) with various types of CHD and 139 age-matched healthy controls (57% females) completed questionnaires assessing HRQoL, PA, social support, significant life events in the past year, education level, civil status, and employment status. Medical variables were retrieved from the patients' hospital records. Patients reported worse physical HRQoL than controls but similar mental HRQoL and PA. Female CHD patients showed worse physical and mental HRQoL and poorer PA than males. In CHD patients, a lower educational level and lower physical exercise capacity predicted lower physical HRQoL, but complexity of CHD was not related to HRQoL or PA. Less social support was associated with lower mental HRQoL and poorer PA in CHD patients. Conclusion: Young adults with CHD have impaired physical HRQoL but normal mental HRQoL and PA. Lower physical exercise capacity, female sex, less social support and lower educational level predict an unfavorable quality of life and PA. This subgroup of patients should be monitored more closely and provided with special psychosocial care to improve long-term outcome.

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Abstract

Aims

The aim of this study was to identify medical and psychosocial risk factors for impaired health-related quality of life (HRQoL) and poor psychological adjustment (PA) in young adults with congenital heart disease (CHD).

Methods and Results

A group of 188 patients (43% females, ages 18-30 years) with various types of CHD and 139 age-matched healthy controls (57% females) completed questionnaires assessing HRQoL, PA, social support, significant life events in the past year, education level, civil status, and employment status. Medical variables were retrieved from the patients' hospital records. Patients reported worse physical HRQoL than controls but similar mental HRQoL and PA. Female CHD patients showed worse physical and mental HRQoL and poorer PA than males. In CHD patients, a lower educational level and lower physical exercise capacity predicted lower physical HRQoL, but complexity of CHD was not related to HRQoL or PA. Less social support was associated with lower mental HRQoL and poorer PA in CHD patients.

Conclusion

Young adults with CHD have impaired physical HRQoL but normal mental HRQoL and PA. Lower physical exercise capacity, female sex, less social support and lower educational level predict an unfavorable quality of life and PA. This subgroup of patients should be monitored more closely and provided with special psychosocial care to improve long-term outcome.

Keywords: Congenital heart disease, quality of life, mental health, psychological adjustment

Introduction

Nowadays, the majority of patients born with congenital heart disease (CHD) survive into adulthood. However, patients with severe types of CHD remain at risk for long-term cardiovascular complications and reduced life expectancy.[1] Health-related quality of life (HRQoL) and psychological adjustment (PA) are important parameters of outcome in this novel patient group. HRQoL and PA have been examined in several studies, with mixed results depending on the methodology used and the study population selected.[2–5] Research has shown that medical characteristics alone (e.g., complexity of heart defect, physical exercise capacity) hardly account for the impact of CHD on the daily lives of young adult patients.[6] However, the impact of psychosocial variables (e.g., educational level, adverse life events, social support) on HRQoL and PA in young adulthood has rarely been examined, and findings are inconsistent.[7–11]

Therefore, the aims of this study were twofold. First, we wanted to examine the HRQoL and PA of young adults with CHD and compare it with that of healthy controls. Based on the literature, we hypothesized that patients would have lower physical HRQoL but not more psychological problems. Second, we sought to explore the impact of a wide range of medical and psychosocial factors on HRQoL and PA. We hypothesized that both medical and psychosocial variables would impact HRQoL and PA.

Methods

Participants

Patient group: All young adult patients (18-30 years) with CHD attending our cardiac outpatient clinic between June 2015 and February 2016 were eligible for participation. Of a total of 353 patients, 16 (4.5%) were excluded because of severe neurological impairments, 13 (3.7%) due to comorbid genetic syndromes, and 2 (0.6%) due to severe psychiatric

diagnoses. Additionally, 22 (6.2%) patients had to be excluded because they were not fluent in German. Thus, 300 patients were eligible for inclusion and were asked for participation by a member of the study team. Sixty-five (22%) patients declined participation, and 47 (15%) consented but did not return the questionnaires. Thus, the final sample comprised 188 participating patients (response rate 63%). The 112 patients who did not fill in the questionnaire had signed the general consent for research, which allowed e use of their medical data for research purposes. These patients are referred to below as non-participating patients.

Control group: Control subjects were 139 sex- and age-matched healthy peers of the patients. Possible controls were identified by the participating patients.

Procedure

The study was approved by the local ethics committee and conducted in accordance with the Declaration of Helsinki. Eligible patients were informed about the study during an outpatient consultation. If they had no scheduled consultation during the study period, they were contacted by phone or e-mail. When written informed consent was given, the questionnaires were handed out directly or sent by post. To select healthy controls, the patients were given a study information sheet and a questionnaire to forward to a good friend of the same sex. The control peers who participated in the study (n = 139) completed and returned the questionnaires along with their signed informed consent. To encourage a higher response rate, participants who had not returned their questionnaires after two weeks were reminded by phone or e-mail to send back the material.

Measures

Medical variables: The following medical variables were retrieved from the patients' hospital records: Type of CHD, maximum physical exercise capacity defined as relative VO_2 max, systemic ventricular ejection fraction, previous cardiovascular complications (e. g. myocardial infarction, heart failure, stroke, endocarditis, atrial fibrillation, supraventricular tachycardia, ventricular tachycardia, third-degree atrioventricular block), type and number of previous surgical and interventional procedures, and number of open heart surgeries performed with cardiopulmonary bypass.

Type of CHD was classified as simple, moderate, or complex defects, as previously proposed.[12] Physical exercise capacity was assessed with symptom-limited cardiopulmonary-exercise testing on a bicycle ergometer. Absolute maximum oxygen uptake (VO_2max) was measured continuously. The individual VO_2max was normalized for age, sex, and height and calculated as percentage of the predicted exercise capacity. This relative VO_2max was classified as severely reduced exercise capacity (< 60%), reduced exercise capacity (60 – 85%), or normal exercise capacity (> 85%). Ventricular function was determined by echocardiography (n=170) or by cardiac magnetic resonance imaging (n=18). Subaortic ventricular dysfunction was categorized into three groups: no subaortic ventricular dysfunction (ejection fraction >52%), mild subaortic ventricular dysfunction (ejection fraction 40 - 52%), and moderate or severe subaortic ventricular dysfunction (ejection fraction <40%).

Health-related quality of life: Self-reported HRQoL was assessed using the German version of the Medical Outcomes Study Short Form-36 item questionnaire (SF-36).[13] The SF-36 is a widely used, generic, and multidimensional measure of HRQoL that assesses eight domains: physical functioning, role limitations due to physical problems, bodily pain, general health perception, mental health, role limitations due to emotional problems, vitality, and social functioning. Each subscale is scored from 1 to 100, with higher scores indicating better

HRQoL. The subscales can be summarized into a physical component summary score (PCS) and a mental component summary score (MCS), which are standardized with a mean of 50 (SD = 10). The SF-36 has consistently been reported to have excellent psychometric properties.[14] We used the questionnaire with a time frame of 4 weeks before assessment.[15] In the current study, the internal consistencies of the PCS and the MCS were good, with Cronbach's α = .78 and α = .77, respectively.

Psychological adjustment: PA was assessed with the German version of the short form (SCL-27) of the Symptom Checklist SCL-90-R.[16] The SCL-27 is a well validated multidimensional questionnaire for evaluating the severity of psychopathological symptoms in the preceding four weeks. Items were rated on a 5-point Likert scale, and the global severity index (GSI) was used as measure of overall PA, with higher scores indicating poorer mental health. In the current study, the internal consistency of the GSI was excellent (Cronbach's α = .91).

Social support: Self-reported social support was determined by the short form (F-SozU-K-14) of the Social support Questionnaire (F-SozU).[17] The 14 items are rated on a 5-point Likert scale and measure several dimensions of subjectively perceived social support, such as emotional and practical support and social integration. We used the sum score (range = 14-70) in our analyses, with higher values reflecting greater social support. The reliability and validity of the F-SozU-14 are well-documented.[17] In the current study, the internal consistency of the scale was excellent (Cronbach's α = .91).

Life events: The occurrence of 12 significant life events in the past year, such as unemployment, birth of a child, or death of a loved one was determined by means of the Life Event Scale (Landolt, unpublished manuscript). A sum score was computed of the number of reported life events for each participant.

Sociodemographic variables: Educational level, civil status, and employment status were assessed. Educational level was measured with reference to the Swiss education system using a scale ranging from 1 to 5 points, with higher scores indicating a higher level.

Statistical analyses

Data were analyzed using IBM SPSS statistical software for Windows, release 22. Student t-tests and χ^2-square tests were used to compare medical and sociodemographic characteristics between participating and non-participating patients to examine whether there was any selection bias. Differences between patients and controls were examined using independent Student t-tests, χ^2-tests, Fisher's exact test, and Mann-Whitney U tests as appropriate. Kolmogorov-Smirnov tests revealed non-normal distributions for the SF-36 and the SCL-27 subscales. Thus, Mann-Whitney U tests for independent samples were used for the between-group analyses of patients and control subjects. Effect sizes were calculated using Cohen's d to express the size of group differences.[18]

For multivariate analyses the PCS, the MCS and the GSI were dichotomized. MCS and PCS values below the first quartile were defined as low physical and low mental HRQoL. PA scores over the third quartile were defined as poor. Three logistic regression models were performed with these three dichotomized scores as dependent variables. Selection of predictors was based on statistical significance of bivariate correlations and previous findings in this field of research and included the following variables: sex, age, educational level, civil status, employment status, complexity of heart defect, number of cardiac interventions in adulthood, number of cardiopulmonary bypass surgeries, current maximum physical exercise capacity, social support, and number of significant life events. The variables civil status and employment status were dichotomized into single versus in a relationship/married, and unemployed versus employed, respectively. Medical interventions in adulthood (age > 18

years) and number of cardiopulmonary bypass surgeries were dichotomized as no versus at least one surgery or heart catheter intervention, and no versus at least one cardiopulmonary bypass surgery, respectively.

Results

Sample characteristics

Sociodemographic and medical characteristics of participating and non-participating patients are provided in Table 1. The two groups did not differ significantly in age, sex, severity of CHD, or systemic ventricular function. However, participants tended to have better physical exercise capacity than non-participants. More males than females participated in the study (p=.049). However, female and male patients were equivalently distributed across types of CHD (χ^2=1.66, p=0.44). Comparison of participating patients (n=188) with controls (n=139) showed no differences in sociodemographic characteristics (Table 2). Diagnoses and classification of CHD based on disease severity are shown in supplemental Table 3.

Health-related quality of life

Comparison of SF-36 domain scores between patients and controls demonstrated significantly worse physical functioning and lower general health perception in patients (Table 4). In addition, the physical component summary score was significantly lower in the patient group. Effect sizes were medium to large. All other scales were similar between patients and controls.

Psychological adjustment

As Table 4 shows, CHD patients and controls showed similar levels of psychological adjustment in all domains of the SCL-27.

Predictors of health-related quality of life and psychological adjustment

The results of logistic regression analyses predicting low HRQoL and poor PA are shown in Table 5. Risk factors for low physical HRQoL were female sex, low educational level, and limited physical exercise capacity. The odds for impaired mental HRQoL were significantly higher for females and patients reporting less social support. Risk factors for poor PA were female sex, lower perceived social support, and a higher number of stressful life events in the past year. Notably, medical variables such as complexity of CHD, number of previous interventions, and occurrence of previous complications did not predict HRQoL or PA.

Discussion

This study demonstrates generally good HRQoL in young adults with CHD, although lower than in the matched healthy peer control group in their general health perception and overall physical HRQoL. Importantly, no differences were found between patients and healthy peers in either mental HRQoL or PA. Our results are therefore in line with our hypotheses and consistent with the majority of previous studies.[2,8,19–22] Previous research showing contrasting results lacked peer control groups, which may explain the inconsistency in results.[8,19] We believe that the use of a peer-control-group instead of community norms allowed for a better control of socio-demographic factors, such as educational level and socioeconomic status.

Our results are very encouraging because they suggest that CHD does not necessarily lead to reduced mental HRQoL or PA in young adulthood when good cardiological care is provided. Researchers studying patients with other chronic diseases have explained this finding with the so-called response shift.[23] This theory proposes that chronically ill patients adapt to their disease by changing internal standards and values and thus can maintain good HRQoL and psychological health despite physical or functional impairments.

This study also analyzed the extent to which medical, psychosocial, and demographic variables were associated with low HRQoL and poor PA. Multivariate analyses revealed that lower physical HRQoL was associated with lower physical exercise capacity, lower educational level, and female sex.

The associations between lower exercise capacity and lower physical HRQoL have previously been found in adults with CHD.[21] The effect of exercise capacity on HRQoL might be underestimated in the current study, because participating patients tended to have a higher exercise capacity than non-participating patients (Table 1). Compared to maximal exercise capacity, submaximal exercise capacity may be even more important in day-to-day live and should be explored for its association with quality of live in future studies.

Our finding that lower educational level is associated with lower physical HRQoL supports previous findings,[7] but the direction of causality remains unclear. Higher education might facilitate understanding of the illness and lead to easier access to medical care, thus possibly leading to better adaptation to the illness. Conversely, patients with lower physical HRQoL could experience more problems during education and as a result achieve a lower educational level.

Our finding that female sex predicted lower physical HRQoL is in line with some previous results[9] but contrasts with another study in young adults with CHD.[2] However, that study, which found no association between physical HRQoL and sex[2], used a short form of the SF-36, which might not be sensitive enough to detect sex differences. Notably, our results are closely in line with epidemiological studies in the general population, where women tend to report lower physical HRQoL than men when assessed with the SF-36 questionnaire.[13,24] Therefore, the sex difference in our CHD patients may not be related to the CHD but might rather be a general phenomenon. This assumption is supported by additional analyses of our data that did not show any significant differences regarding physical HRQoL between males

and females, neither in patients with simple, moderate nor in patients with complex CHD (data not shown).

As in the present study, female sex has previously also been reported to be associated with lower mental HRQoL[7,9] and poor PA.[10] Similar to physical HRQoL, this finding is well known from epidemiological studies in the general population, where women consistently report lower mental HRQoL and higher rates of mental health problems than men.[13,24,25] However, recent results of studies that assessed PA with the SCL-27 indicated no sex differences in PA except for the 'vegetative symptoms' scale.[26,27] Thus, it remains unclear whether the poorer PA in our female patients reflects the situation of the general population or is associated with the CHD in some unknown way. However, although most women with repaired congenital heart defects will tolerate pregnancy, the risk of adverse events during pregnancy is increased in almost all women and there is in addition an increased risk of recurrence in the offspring. In contrast to the pediatric setting, pregnancy risks and issues around contraception are routinely discussed during outpatient visits in the adult CHD clinics and thus may raise the awareness of affected women for their heart disease, even though most adolescents and young adults with congenital heart disease are asymptomatic. In our experience, this awareness is typically much more pronounced in female patients compared to their male counterparts of the same age. This may therefore be one possible explanation for the findings in our study but certainly needs further exploration in future studies. Interestingly, additional analyses revealed that only female patients with simple, but not with moderate or complex CHD reported impaired PA compared with their male counterparts. Furthermore, only female CHD patients with moderate, but not with simple or complex CHD reported lower mental HRQoL than male patients. Further studies are needed to explore this association between gender differences regarding HRQoL and PA and complexity of CHD.

In accordance with our results, social support has previously been shown to contribute to a better mental HRQoL[8,9] and better PA.[10,11] Social support may help patients to cope better with their CHD disease and thus may lead to a better adaptation to the disease. The finding that a higher number of critical life events was associated with poorer PA in CHD patients has not been reported before. It is clear that non-CHD-related adverse life events can compromise adaptation to the disease and therefore need to be taken into account in follow-up care.

Surprisingly, medical variables such as CHD complexity, number of previous interventions, and medical complications had no significance association with either HRQoL or PA. This finding contrasts with a meta-analysis of SF-36 data, which found lower physical HRQoL in moderate or complex CHD than in simple CHD.[28] However, the results of the meta-analysis could have been influenced by differing methodological approaches (type of CHD included, categorization of CHD complexity) and the inclusion of broader age groups. In line with our results, several studies have shown that the complexity of CHD is unrelated to PA.[11,22] Our findings suggest that, at least in the Swiss population of young adults with CHD, a patient's current physical exercise capacity might indicate physical HRQoL better than the disease severity, possibly because exercise capacity better reflects the actual condition of the patient than the initial classification of the CHD. One may also speculate that many young adults with CHD are not yet aware of their limitations and their long-term risk of medical complications. Thus, complexity of CHD might play a more important role in HRQoL and PA in middle-aged patients.

Limitations

This study has several limitations which need to be kept in mind when interpreting the results. First, the cross-sectional study design does not allow any causal conclusions to be drawn.

Second, because patients who were not fluent in German had to be excluded from the study, our results may not be representative of patients with migration background. Furthermore, patients with a genetic syndrome as well as other congenital or acquired neurological impairments leading to mental disorders were excluded from the study. Thus, our results may not be representative for this specific subgroup of young adults with CHD. Third, the participating patients tended to have better exercise capacity than non-participating patients. Fourth, given that treatment for hypoplastic left heart syndrome at our center was offered in early 2000s, the cohort of adult survivors with hypoplastic left heart syndrome, a particularly vulnerable group of adult Fontan-patients is certainly underrepresented. Given their high risk of complications as young adults, these patients may experience worse quality compared to other Fontan patients[29]. Fifth, the SF-36 questionnaire is a generic measure of HRQoL and so may lack sensitivity for the specific problems of patients with CHD. However, it has been used in many previous studies and is a well validated and widely accepted measure.

Strengths of the present study are a matched healthy peer control group, something often lacking from previous studies. Furthermore, our large sample size allowed a high statistical power of over 0.95 (with an alpha error of .05). Additionally, socio-demographic and psychosocial variables were assessed with standardized questionnaires, which allowed a comprehensive overview of HRQoL and PA and its predictors.

Implications

Several implications for clinical management and future research activities can be drawn from our findings. Importantly, our results suggest that, independently of the complexity of the heart defect, specific subgroups of young adults with CHD are at risk of an unfavorable outcome, particularly female patients and patients with lower exercise capacity, lower educational level and less social support. These patients should be monitored more closely,

and screening instruments for assessing HRQoL and psychological well-being should be implemented into routine medical care. Appropriate psychosocial care should be provided to patients at risk to improve long-term outcome.

From a methodological perspective, longitudinal data are needed to establish causal links between medical and psychosocial variables and HRQoL and PA. Moreover, further studies should investigate whether and how HRQoL and PA change over the course of the disease.

Conclusions

HRQoL and PA in this Swiss cohort of young adults with CHD was generally good and comparable with that of healthy peers, with exception of some dimensions of physical HRQoL. However, lower exercise capacity, female sex, lower social support, and lower educational level were identified as risk factors for unfavorable outcome. At-risk patients should be monitored closely and if needed, provided with appropriate interventions.

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Conflict of interest

None declared

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Table 1 Patients' characteristics

	Participants (*n*=188)		Non-participants (*n*=112)		*p*
Age (years)					0.71[a]
Mean (SD)	24.7	(3.7)	24.5	(3.8)	
Sex					0.31[b]
Female	80	(42.6%)	41	(36.6%)	
Male	108	(57.4%)	71	(63.4%)	
Complexity of CHD					0.74[b]
Simple	61	(32.4%)	41	(36.6%)	
Moderate	74	(39.4%)	40	(35.7%)	
Complex	53	(28.2%)	31	(27.7%)	
VO_{2max} predicted[c]					0.07[a]
Mean (SD)[d]	96.3%	(21.7%)	91.1%	(23.1%)	
Normal exercise capacity	129	(68.6%)	57	(50.9%)	
Reduced exercise capacity	44	(23.4%)	37	(33.0%)	
Severely reduced	7	(3.7%)	5	(4.5%)	
Systemic ventricular dysfunction					0.37[b]
No ventricular dysfunction	158	(84.0%)	100	(89.3%)	
Mild ventricular dysfunction	27	(14.4%)	10	(8.9%)	
Moderate or severe ventricular dysfunction	3	(1.6%)	2	(1.8%)	

[a]t test for independent samples.
[b]Pearson chi-square test (χ^2).
[c]Maximum physical capacity
[d]Due to missing values, the sample size for this analysis was reduced to *n*=180 and *n*=99 for the participants and non-participants, respectively.

Table 2 Comparison of patients and healthy control subjects

	Patients (*n*=188)	Controls (*n*=139)	*P*-value
Age (years)			0.25[a]
Mean (SD)	24.7 (3.7)	24.2 (3.7)	
Sex			0.91[b]
Female	80 (42.6%)	60 (56.8%)	
Male	108 (57.4%)	79 (43.2%)	
Swiss nationality	157 (83.5%)	120 (86.3%)	0.14[c]
Educational level			0.46[a]
Mandatory school without certificate	4 (2.1%)	0 (0.0%)	
Completed apprenticeship	93 (49.5%)	68 (48.9%)	
A-level equivalent	20 (10.6%)	16 (11.5%)	
College of higher education	59 (31.4%)	43 (30.9%)	
University	12 (6.4%)	12 (8.6%)	
Number of significant life events in the past year[e]			0.26[d]
Mean (SD)	2.2 (1.7)	1.9 (1.5)	
None	30 (16.0%)	29 (21.0%)	
1 Significant life event	40 (21.3%)	27 (19.6%)	
2 Significant life events	52 (27.7%)	41 (29.7%)	
≥ 3 Significant life events	66 (35.0%)	41 (29.7%)	
Social support			0.95[d]
Mean (SD)	62.2 (7.3)	62.3 (7.4)	

[a]t test for independent samples.
[b]Pearson chi-square test (χ^2).
[c]Fisher's exact t-test
[d]Mann-Whitney U Test for independent samples.
[e]Due to missing values, the sample size for this analysis was reduced to *n*=186 and *n*=138 for the patients and control subjects, respectively.

Table 4 Comparison of health-related quality of life of patients and control subjects

	Patients (*n*=188)		Controls (*n*=139)		Mann-Whitney U-Test	Effect size
	M	*SD*	*M*	*SD*	*p*	*d*
SF-36 scales						
Physical functions	93.1	10.7	97.5	6.6	**<.001**	0.49
Role physical	93.3	19.7	94.4	19.5	.25	0.07
Bodily pain	91.4	18.7	89.8	17.9	.16	- 0.09
General health	73.3	20.1	82.0	14.2	**<.001**	0.50
Vitality	60.5	17.3	60.0	16.4	.57	- 0.03
Social functions	91.0	17.1	92.8	12.9	.65	0.11
Role emotional[a]	87.3	26.3	89.2	23.8	.60	0.07
Mental health	76.2	14.6	75.4	14.1	.48	- 0.05
Health transition[a]	2.7	0.7	2.7	0.7	.60	-0.03
Physical component summary[a]	54.6	6.7	56.3	5.0	**<.001**	0.30
Mental component summary[a]	50.0	8.4	49.7	7.7	.37	- 0.03
SCL-27 scales						
Global Severity Index	0.5	0.4	0.4	0.4	.25	-0.11
Depressive symptoms	0.4	0.5	0.3	0.4	.35	-0.17
Dysthymic symptoms	0.6	0.6	0.6	0.6	.54	-0.04
Vegetative symptoms	0.4	0.4	0.4	0.5	.11	-0.09
Agoraphobic symptoms	0.2	0.3	0.2	0.3	.80	-0.02
Sociophobic symptoms	0.5	0.6	0.5	0.6	.78	-0.00
Symptoms of mistrust	0.6	0.6	0.5	0.6	.16	-0.18

Effect Size according to Cohen: 0.20, small effect size, 0.50, medium effect size, >0.80 large effect size.

[a]Due to missing values, the sample size for the patient sample was reduced to *n*=187.

Table 5 Summary of logistic regression analyses predicting low physical HRQoL (*n*=177), low mental HRQoL (*n*=177) and impaired PA (*n*=178)

	Low physical HRQoL					Low mental HRQoL					Impaired PA				
Variables	B	SE	Odds ratio	95% Confidence Interval	Wald statistic	B	SE	Odds ratio	95% Confidence Interval	Wald statistic	B	SE	Odds ratio	95% Confidence Interval	Wald statistic
Female sex	1.43	0.44	4.19	1.77–9.90	10.68**	0.86	0.42	2.37	1.05–5.38	4.28*	1.30	0.45	3.66	1.50–8.95	8.14**
Age	0.00	0.06	1.00	0.89–1.14	0.00	0.00	0.06	1.00	0.88–1.13	0.00	-0.04	0.07	0.96	0.84–1.10	0.31
Educational level	-0.47	0.23	0.62	0.40–0.97	4.33*	-0.03	0.20	0.97	0.65–1.43	0.02	0.30	0.21	1.35	0.89–2.04	2.04
Civil status[a]	-0.10	0.47	0.91	0.36–2.27	0.04	-0.46	0.46	0.63	0.26–1.54	1.03	-0.41	0.48	0.67	0.26–1.69	0.73
Employment status[b]	-0.40	0.53	0.67	0.24–1.90	0.56	0.00	0.48	1.00	0.38–2.58	0.00	0.66	0.52	1.93	0.70–5.33	1.60
Complexity of heart defect	-0.15	0.31	0.86	0.47–1.57	0.24	-0.17	0.29	0.84	0.48–1.48	0.36	-0.14	0.31	0.87	0.48–1.60	0.20
Medical interventions in adulthood[c]	0.63	0.57	1.87	0.61–5.77	1.19	0.29	0.54	1.34	0.46–3.91	0.29	0.86	0.55	2.37	0.81–6.96	2.46
Number of HLM interventions[d]	-0.15	0.27	0.86	0.50–1.46	0.31	-0.36	0.26	0.70	0.42–1-16	1.91	-0.14	0.26	0.87	0.52–1.46	0.27
Maximum physical capacity (%)	-0.04	0.01	0.96	0.94–0.98	10.10**	0.00	0.01	1.00	0.98–1.02	0.01	-0.01	0.01	0.99	0.97–1.01	0.31
Social support	0.00	0.03	0.99	0.94–1.05	0.03	-0.11	0.03	0.90	0.84–0.94	16.52***	-0.14	0.03	0.87	0.82–0.92	20.52***
Number of SLE in the past year	0.04	0.13	1.04	0.81–1.34	0.12	0.16	0.12	1.17	0.92–1.50	1.67	0.31	0.13	1.36	1.05–1.76	5.56*

HLM, heartlung machine; SLE, significant life events

* $p < 0.05$, ** $p < 0.01$, *** $p < 0.001$

[a]1=single, 2=in a relationship/married/concubinage

[b]1=unemployed, 2=employed

[c]0=no medical interventions in adulthood, 1=at least one surgery or/and heart catheter intervention in adulthood

[d]0= no HLM surgery, 1= at least one HLM surgery

Table 3 Distribution of congenital heart defects stratified for disease complexity according to the guidelines of Warnes et al., 2001

Simple CHD	*n* (%)	Moderate CHD	*n* (%)	Complex CHD	*n* (%)
Bicuspid aortic valve	23 (12.2)	Coarctation of the aorta	29 (15.4)	Pulmonary atresia	16 (8.5)
Repaired Ventricular septal defect	11 (5.9)	Ebstein's anomaly	18 (9.6)	Fontan procedure	14 (7.4)
Isolated mitral valve disease	11 (5.9)	Infundibular pulmonary stenosis	8 (4.3)	TGA[a] - atrial switch (Senning, Mustard)	11 (5.9)
Isolated Pulmonary valve stenosis	4 (2.1)	Atrioventricular septal defect	7 (3.7)	TGA[a] - arterial switch	5 (2.7)
Isolated aortic valve disease	3 (1.6)	Subvalvular aortic stenosis	3 (1.6)	TGA[a] - congenitally corrected	3 (1.6)
Atrial septal defect II - closed	2 (1.1)	HCM[b] / HOCM[c]	3 (1.6)	Other complex CHD[e]	2 (1.1)
Small Ventricular septal defect	2 (1.1)	Tetralogy of fallot	2 (1.1)	Eisenmenger syndrome	1 (0.5)
Sinus venosus defect - closed	2 (1.1)	Double outlet right ventricle (DORV)	1 (0.5)	Cyanotic heart disease[f]	1 (0.5)
Other simple CHD	2 (1.1)	Marfan syndrome	1 (0.5)		
Small patent ductus arteriosus	1 (0.5)	Shone's complex	1 (0.5)		
		Right ventricular dysplasia	1 (0.5)		
Total	61 (32.4)		74 (39.4)		53 (28.2)

[a]Transposition of the great arteries
[b]Hypertrophic cardiomyopathy
[c]Hypertrophic obstructive cardiomyopathy